

Breast Reconstruction by Latissimus Dorsi Myocutaneous Flap and Silicone Implant in a Patient with Poland's Syndrome: A Case Report

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Case Report

Abstract:

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Background: Poland's syndrome is an uncommon congenital abnormality characterized by unilateral lack of the pectoralis major muscle, ipsilateral brachysyndactyly, and is occasionally linked with other anterior chest wall deformities. **Case Report:** A 28-year-old woman with marked hypoplasia of the right breast without any malformation of the right upper limb since birth was studied. Reconstruction was done with pedicled Latissimus Dorsi myocutaneous flap and silicone implant along with contralateral mastopexy to manage this case. The current literature on the topic is also reviewed. **Conclusion:** The surgical method with combined autologous pedicled Latissimus Dorsi myocutaneous flap with implant is a reliable technique for breast reconstruction and can deliver an excellent outcome in long term in selected cases of Poland's syndrome.

Keywords: Poland's Syndrome, Breast reconstruction, Latissimus Dorsi flap, Breast implant.

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INTRODUCTION

Poland's syndrome was first described by Sir Alfred Poland in 1851 which is a rare congenital malformation of the chest wall and upper limb in the form of absence of the pectoralis muscles and ipsilateral hand abnormality with an incidence of 1 in 7000 to 100,000.¹ It has a gender predominance in men at a ratio of 3:1 with 60% to 75% affecting the right side². It usually presents as unilateral, bilateral anomalies also have been found. Most cases are sporadic but familial cases have been also reported.² Classically, Poland's syndrome is defined as hypoplasia or lack of the sternocostal head of the pectoralis major, which results in the absence of the anterior axillary fold, subclavicular hollowing and a pathognomonic groove at the junction of the superior anterior axillary line and chest wall. However, recent definition is that Poland's syndrome comprises hypoplasia/absence of the pectoralis major with at least two of these minor criteria: hypoplasia or absence of the breast, absence of the nipple, absence of axillary hair, absence of adjacent muscles (e.g., latissimus dorsi, serratus anterior, external oblique, deltoid, infraspinatus, supraspinatus, and pectoralis minor), absence of costal cartilage and anterior ribs, absent subcutaneous tissue, axillary webbing, ipsilateral brachydactyly, brachysyndactyly or amelia.³

It is quite difficult to classify Poland's syndrome clinically because of the diversity in presentation. The general classification used was introduced by Foucras et al. and categorized the severity of disease as mild, moderate and severe.⁴ Breast anomalies among females are variable and range from mild hypoplasia to amastia. Poland's syndrome is involved in 14% of breast aplasia.

In the mild form, structural abnormalities may only be detected radiographically. These patients may be referred to the plastic surgeon for mild asymmetry, without any formal diagnosis. The mild variant of Poland's syndrome is more common than the classic full presentation with an incidence of 1 in 16,500 live births. The moderate variant of Poland's syndrome represents the classic form and is characterized by hypoplasia of breast parenchyma, high IMF, an underdeveloped and superiorly displaced NAC, with the absence of the anterior axillary fold. The severe variant of Poland's syndrome represents the most challenging to reconstruct and is characterized with a marked deformity of the chest wall with tight chest skin and axillary webbing which might require reconstruction of thoracic cage.

CASE REPORT

A twenty-eight years old married female, mother of one presented to Sheikh Hasina National Institute of Burn and Plastic Surgery with right hypoplastic breast for reconstruction. She was born with Poland's syndrome of moderate variant with absence of pectoralis major with hypoplastic breast which was more visualized during her adolescent period. She also had chest wall deformity (deficient 2nd, 3rd costal cartilage), loss of anterior axillary fold and groove at the junction of anterior axillary line and chest wall, deficient skin and soft tissue, ill-defined IMF, hypoplastic areola with NAC placed much superiorly in comparison with the left breast. She had no deformity of upper limb. She was within normal limits in height, weight and intelligence.

Patient had multiple abscesses under her right axilla followed by incision and drainage and later closure done at multiple sites at the age of 8 months which left her with widespread multiple scars around right axilla.

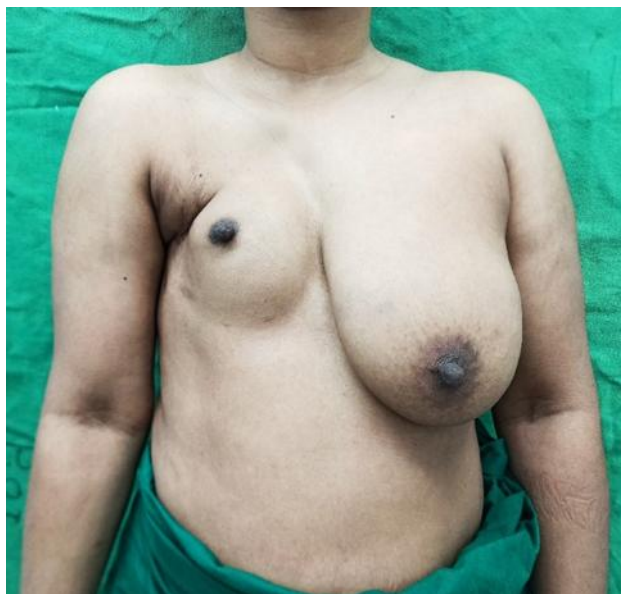


Fig 1: Young lady with Poland's Syndrome

Reconstruction of right hypoplastic breast was performed under general anaesthesia with pedicled Latissimus Dorsi (LD) myocutaneous flap and placement of implant under the muscle. Marking of desired breast footprint and optimal positioning of the flap were drawn preoperatively on the patient in standing position. Ipsilateral LD myocutaneous flap was raised in lateral position and delivered through a tunnel and inset done at upper outer quadrant of the breast.



Fig 2: Markings for LD Flap harvest on right side along with symmetrizing mastopexy on left side

The LD muscle was harvested as large as possible and muscle edges were sutured with chest wall.

A round, textured silicone implant of 260cc was inserted in subcutaneous plane and covered by LD myocutaneous flap. 3-0 vicryl suture was used to fix the flap. The inferior margin of the implant was placed at the marked inframammary fold.



Figure 3: Perioperative picture of LD myocutaneous flap harvest A small suction drain was placed alongside the implant



Fig 4: Before and after LD flap with implant along with contralateral symmetrization surgery (14th POD)

She had grade 2 ptosis of left breast and her cup size was C. So, contralateral mastopexy was performed for symmetrization at the same setting. The patient had an uneventful postoperative course. Patient was on follow up and is highly satisfied with the results. Although we recommended her further revision surgery for more natural and aesthetic outcome, she is not interested to go ahead with any further procedures.



Fig 5: Before and after surgery - same patient in right oblique view (8 months follow up)

DISCUSSION

Poland's syndrome is a rare combination of chest wall, breast and hand deformities and selecting the appropriate treatment is challenging for reconstructive surgeons. Surgeons must provide a wide variety of treatment choices, and a mix of diverse surgical procedures is frequently appropriate for patients to obtain the desired outcome. Surgeons must provide a wide variety of treatment choices, and a mix of diverse surgical procedures is frequently appropriate for patients to obtain the desired outcome.² The repair may be done in one or two stages according to severity. Treatment option ranges from lipo transfer, autologous flap transfer, augmentation by implants with or without pre-expansion and free flaps depending on presentation. In case of severe variant of chest wall deformity or hand deformity further reconstruction is necessary.

Male patients with mild variety of Poland's syndrome can easily be managed with autologous lipo transfer. In children with mild to moderate forms of Poland's syndrome, which is limited to absence of the pectoralis muscles and breast hypoplasia, the operation should be postponed until after puberty at that time, an LD flap transposition combined with breast augmentation by implant may be carried out in female patients.

LD flap is well established, reliable and most commonly used pedicle flap for breast reconstruction. Its close proximity to the chest wall and breast provides an advantage compared to more complex techniques, such as microsurgical free flap reconstructions and their potential risk for anastomosis related complications. It is a broad muscle and has a good axial rotation that covers a large area and can be used with or without skin paddle for reconstruction of small to moderate breasts. However, the muscle harvested with a skin paddle does not typically provide sufficient volume for large breast reconstruction. To achieve desired breast volume the LD flap is often used in combination with silicone implants which we used in this particular case as myocutaneous flap because of deficient skin and soft tissue.⁵ To obtain perfect nipple symmetry, the muscle was primarily secured to the chest wall for creating a stable base and adequate projection. The implant surface needs to be covered with muscle, which is sometimes not possible due to small muscle length. If the skin cover of an implant is not flexible enough, it will not allow stretching in a single stage and it will be necessary to implement a tissue expander first.

In some cases of Poland's syndrome with absent NAC, the nipple can be simultaneously reconstructed with a thoracodorsal artery perforator (TAP) flap in breast reconstruction by LD myocutaneous flap.⁶ In recent literatures, other techniques, e.g., laparoscopic reconstruction using the omentum flap technique or autologous fat injection, are also described.⁷ Sometimes contralateral mastopexy or reduction mammoplasty may be needed and often desired by the patients for symmetrization.⁶ Postoperative complications are very few following breast reconstructions by LD flap (mostly seroma), which are easy to manage.⁸ A sound and precise surgical technique may deliver very good and satisfactory outcome.

CONCLUSION

Due to the different grades of severity and the complex combination of different deformities, managing patients with Poland's syndrome can be extremely challenging for surgeons. The type of surgery depends on the extent of the malformation and individual patient preferences. A severely hypoplastic breast can benefit with LD muscle or myocutaneous flap along with a breast implant. This type of reconstructive surgery is recommended for aesthetic as well as psychosocial wellbeing of these type of patients.

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